THE NEW ARMENIAN MEDICAL JOURNAL
Vol.16 (2022), No 1, p.29-34
DOI: https://doi.org/10.56936/18290825-2022.16.1-29

CAROTID BODY TUMORS: CASE SERIES OF EXTREMELY RARE HEAD AND NECK PARAGANGLIOMAS

Tajibayev T.K., Chormanov A.T., Matkerimov A.Zh., Tergeussizov A.S., Baubekov A.A., Zhakubayev M.A., Sagatov I.Y*., Kanchi M.

Department of Vascular Surgery, National Scientific Center of Surgery named after A.N.Syzganov, Almaty, Kazakhstan.

Received 23.05.2021; accepted for printing 18.01.2022

Abstract

Carotid body tumors, also commonly known as nonchromaffin paragangliomas and chemodectomas, is a slowly growing neoplasm originating from carotid body chemoreceptors. Herein we have presented a case series of surgical treatment of patients with carotid body tumors in National Scientific Center of Surgery named after AN Syzganov (2009-2020). Total number of patients was 10 with 11 neck mass, of which 7 (70%) were women, the average age was 47 (31-73) years, one case we faced with a bilateral location. The main complaint was slow-growing neck mass. On computed tomography angiography most of cases (70%) were of the 3th type according to the Shamblin grade with average size of 5.9 (4-8) cm. The main treatment was surgical excision in all cases. Mean duration of surgery 111.5 (75-190) min and hospital stay days 12.1(8-20) were registered. Total number of complications were 2 (20%), where in 1 case surgery complicated by bleeding more than 500 ml, and 1 patient presented dysphonia after the surgery, which resolved by time. Duration of surgery and hospital stay days were directly associated with size of mass and distance to base of skull. Pathology results showed that two patients (20%) had malignant cells, one of them with metastases to the nearest lymph nodes. Surgical resection is the treatment of choice for carotid body tumors. The large size of the masses and involvement of the carotid arteries and cranial nerves in the process directly affect the surgical approach and increase the risk of complications.

Keywords: *carotid body tumors, neck paragangliomas, neck mass, nonchromaffin paragangliomas.*

Introduction

Paragangliomas of the head and neck are rare vascular tumors derived from the paraganglia tissues originating from the neural crest and associated with autonomic ganglia along the sympathetic trunk. [*Bougrine F et al., 2008*] Carotid body tumors (CBT), also commonly known as nonchromaffin paragangliomas and chemodectomas, is a slowly growing neoplasm originating from carotid body chemoreceptors [*Pryse-Davies J et al.,1964; Lack E et al., 1977; McPhersen G et al.,1989*]. It occurs in 0.5 - 0.5% of cases of head and neck mass [*Green J et al., 1998; Ceruse P et al., 2014*]. Carotid paraganglioma or carotid body tumors represent 60-70% of paraganglioma of the head and neck. [*Lahey F, Warren K*, 1947; *Parry D et al., 1982; Gerecer-Gil N et al., 2010; Woolen S, Gemmete J, 2016*].

About 1 in 30,000 head and neck formations corresponds to a paraganglioma, 45% of which are CBT. It was found that bilateral location occurs in 5% of sporadic cases, while in family cases the percentage of CBT on both sides reaches 30% [*Power A et al., 2012*]. The frequency of occurrence in women is 2.5-3 times higher than in men

Cite This Article as:
Tajibayev T.K., Chormanov A.T., Matkerimov A.Zh., Tergeussizov A.S., Baubekov A.A., Zhakubayev M.A., Sagatov I.Y., Kanchi M. (2022); Carotid body tumors: case series of extremely rare head and neck paragangliomas.; NAMJ v.16 (2022) No.1, p.29-34; DOI: https://doi.org/10.56936/18290825-2022.16.1-29

Address for Correspondence:
Sagatov I.Y., MD., Ph.D.
JSC "National Scientific Center of Surgery", 62 Zheltoksan street, Almaty 050004, Kazakhstan
Tel: +7 708 155 15 77; E-mail: inkar_sagatov@mail.ru

[*Ward P et al., 1978; Wang S et al., 2000*]. CBT occurs at any age, but is usually diagnosed between the third and sixth decades of life [*Pryse-Davies J et al., 1964*].

In various observations, from 2–9%, the carotid chemodectoma can be malignant with the formation of metastases [*Wang S et al., 2000*]. The presence of metastases to the lymph nodes, as well as the presence of distant metastases, indicates a malignant nature of the tumor and has an unfavorable prognosis [*Mitchell R et al., 1996*].

The first anatomical mention of a carotid glomus tumor was made by Albercht von Haller in 1743 [*Scudder C et al., 1903*]. The first attempt at surgical removal of a chemodectoma was made by Reigner in 1880, which culminated in the death of the patient [*Byrne J et al., 1958*]. In 1886, Maydl first performed a successful operation to remove a chemodectoma, with a subsequent complication in the form of aphasia and hemiparesis [*Scudder C et al., 1903*]. The first operation to remove the tumor with the preservation of the internal carotid artery and without the development of complications was performed in the USA by Scudder in 1903 [*Scudder C et al., 1903*]. Since 1970, with the development of cardiovascular surgery technologies, the widespread introduction of neuroimaging methods, the mortality rate due to surgical removal of a chemodectoma has been less than 10% [*Shamblin W et al., 1971; Robison J et al., 1986*].

Many imaging techniques are available for assessing paraganglioma. Treatment is primarily based on surgical removal. A significant role in the choice of surgical treatment approaches was played by a study by a group of authors Shamblin et al, who proposed their CBT classification based on carotid artery involvement. The first group is CBT without involvement of the carotid arteries, in this position it is easy to remove the tumor from the vessels, the second group has partial involvement of the vessels, in this category partial excision of external layer of the artery is possible, the third group has complete involvement of the vessels and transmural invasion of the tumor, in this case, tumor resection is required along with the carotid arteries due to the inability to separate them from each other [*Williams M et al., 1992*].

To overcome it is possible, due to the uniting the knowledge and will of all doctors in the world

Despite the fact that monitoring of undesirable consequences during surgery, improvement of surgical technique and postoperative monitoring yielded results and reduced the risk of mortality closer to zero, the percentage of postoperative neurological complications remains high and reaches from 10 to 40%. However, due to the locally invasive nature of CBT, the surgical approach is still effective [*Nora J et al., 1988; Daudi F et al., 1989; Bemard R et al., 1992*]. Along with surgical treatment in the literature, there are publications by authors suggesting transarterial embolization (TAE) in the preoperative period, which will reduce the risk of complications [*Muhm M et al., 1997; Li J et al., 2010*]. Herein we have presented a series of 10 cases of surgical treatment of patients with CBT.

MATERIALS AND METHODS

10 patients with 11 neck masses treated by open surgery over the past 10 years (from 2009 to 2021) were retrospectively studied in the National Scientific Center of Surgery named after AN Syzganov. Furthermore, epidemiological, clinical, imaging and surgical features of this rare disease will be discussed. Collection of all patient data, statistical calculations were performed using Microsoft Excel.

RESULTS

The total number of patients was 10, of which 7 (70%) were women, the average age was 47 (31-73) years, one case we faced with a bilateral location. The main complaint was slow-growing neck mass (Table 1). Painful of the neck mass in 1, dysphonia and headache in 1 case and Horner's syndrome in 1 patient were noted in medical history. All the significant clinical presentation was associated with large size of mass. Genetic tests for the succinate dehydrogenase gene mutation have not been conducted, but in one case was familial CBT, which was associated with bilateral location. In 1 case mild carotid artery compression was revealed by ultrasound imagine. On CTA most of cases (70%) were of the 3th type according to the Shamblin grade with average size of 5.9 (4-8) *cm*.

Table 1

Main features (Part 1)

No	Sex	Age, (years)	Clinical presentation	Family history	Side	Classification by Shamblin	Size, (*cm*)
1	M	54	Slow-growing neck mass + Dysphonia + Headache	nonfamilial	right	Type 3	7.6
2	F	48	Slow-growing neck mass+dizziness	nonfamilial	left	Type 3	5.5
3	F	73	Slow-growing neck mass + painfulness	nonfamilial	left	Type 2	5
4	F	43	Slow-growing neck mass	nonfamilial	right	Type 3	6
5	F	38	Slow-growing neck mass+dizziness	Familial	multiply	Type 3	2.5; 8
6	F	33	Slow-growing neck mass + Horner's syndrome+dizziness	nonfamilial	left	Type 3	7
7	M	43	Slow-growing neck mass+dizziness	nonfamilial	right	Type 3	4.5
8	F	31	Slow-growing neck mass+dizziness	nonfamilial	right	Type 2	4
9	F	62	Slow-growing neck mass	nonfamilial	right	Type 3	6.2
10	M	45	Slow-growing neck mass	nonfamilial	left	Type 2	5.3

Table 2

Main features (Part 2)

No	Surgery	Complication	Pathology	Imaging features	Duration of surgery, (min)	Hospital stay, (days)
1	Excision + Ligation of ECA	-	Malignant CBT	-	150	9
2	Excision	-	Non-malignant CBT	-	85	14
3	Excision	Cranial nerve injury	Malignant CBT	-	115	13
4	Excision	-	Non-malignant CBT	-	95	10
5	Excision +ICA replacement	Bleeding	Non-malignant CBT	Mild compression of ICA by US	190	20
6	Excision	-	Non-malignant CBT	-	95	16
7	Excision	-	Non-malignant CBT	-	75	8
8	Excision with ligation of ECA	-	Non-malignant CBT	-	120	11
9	Excision	-	Non-malignant CBT	-	95	11
10	Excision	-	Non-malignant CBT	-	100	9

The main treatment was surgical excision in all cases (Table 2). In 2 cases we used ligation of external carotid artery due to high risk of rupture and in 1 case replacement of internal carotid artery was performed. Mean duration of surgery 111.5 (75-190) *min* and hospital stay days 12.1 (8-20) were registered. Total number of complications were 2 (20%), where in 1 case surgery was complicated by bleeding more than 500 *ml*, and 1 patient presented dysphonia after the surgery, which resolved by time. Duration of surgery and hospital stay days were directly associated with size of mass and distance to base of skull.

Pathology results showed that two patients (20%) had malignant cells, one of them with metastases to the nearest lymph nodes. Patient data were subsequently referred to oncologists and radiation therapy courses were applied to them in the affected area. The average follow-up for these patients was 1 year. There were no other complications such as cerebrovascular disturbances (stroke, transient ischemic attack) and death.

Clinical case

A 31-year-old woman with a slow growing neck mass on the right side and dizziness was admitted to the department of vascular surgery (Fig.

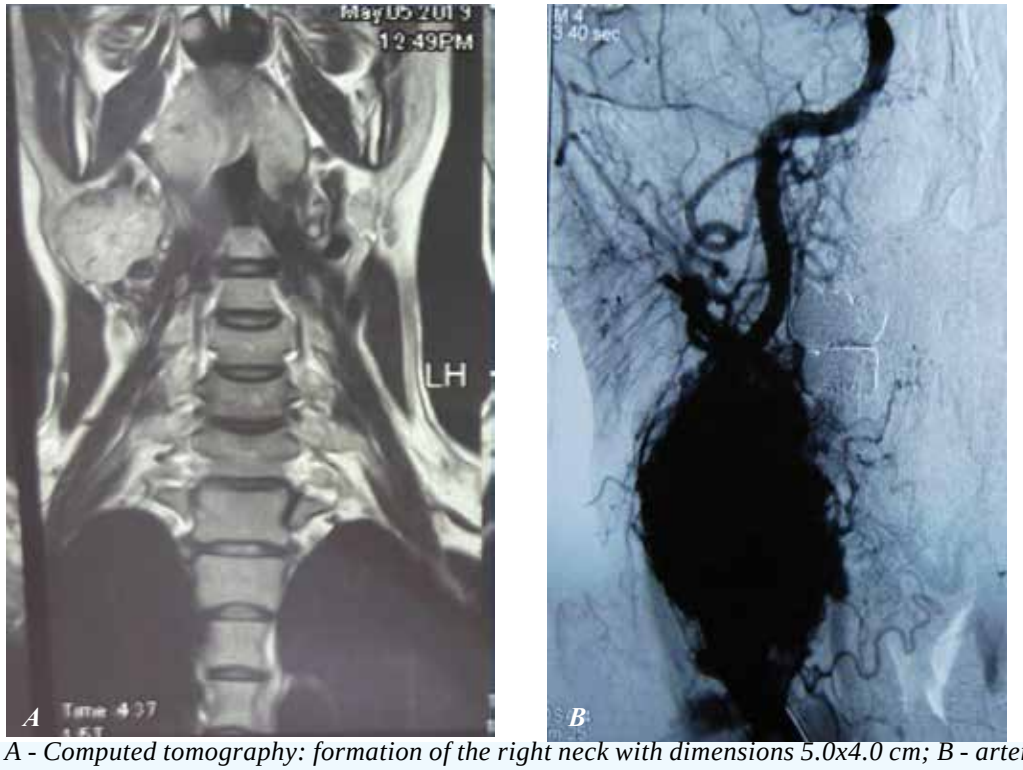

***FIGURE** 1. A - Computed tomography: formation of the right neck with dimensions 5.0x4.0 cm; B - arteriography: there is a hypervascular formation in the area of bifurcation of the right common carotid artery.*

1). After the distribution of patients into groups, a comparative analysis of the perioperative parameters of the patients was carried out. Thus, the average number of days a patient was hospitalized increased in direct proportion to the size and type of tumor.

In addition, we performed several types of surgical treatment (Fig. 2). As shown in the above table, the volume of the operation, the time of the operation, intraoperative blood loss, the volume of blood transfusion increased in the same way as the size of the tumor increased, involvement of the carotid arteries and malignancy of the formation.

DISCUSSION

CBT usually manifests itself as a painless, slowly growing neck formation, laterally to the apex of the hyoid bone [*Werter I et al., 2013*]. Sometimes the patient complains of hoarseness, dysphonia and even less often dizziness and coughing [*Lim J et al., 2010*]. In our study, only one patient had symptoms of grade 1 dysphonia, which was presumably due to the large size of the tumor (8.0 x 6.5 *cm*).

As a rule, the asymptomatic course of the disease in the initial stage and the slow growth of CBT can underestimate the condition and late

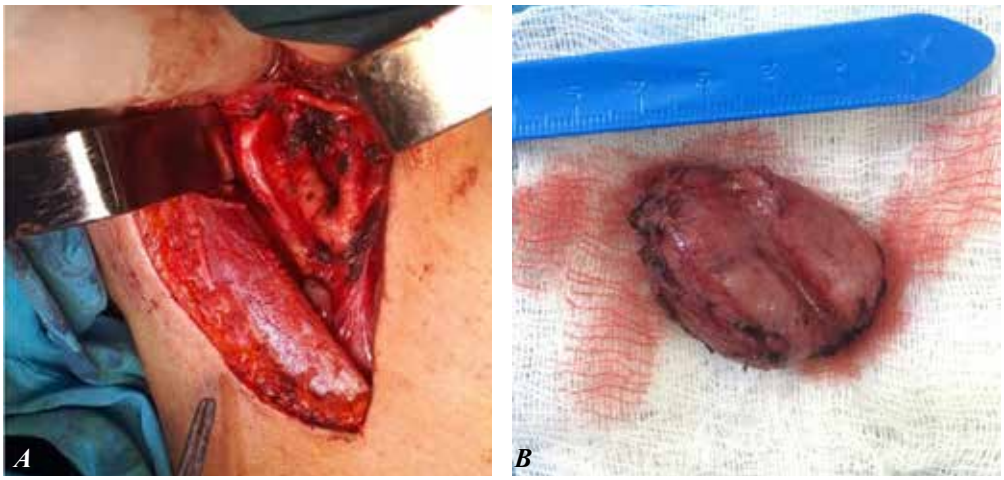

***FIGURE** 2. A - Intraoperative view of the carotid bifurcation after removal of CBT; B - View of formation after excision.*

treatment of patients. So, in our center there was only one case with the first type of tumor according to the Shamblin classification, while the third type was in half of the patients, although many of them noted the presence of a tumor-like formation in the neck area was sick for 10 years. Moreover, in 5 out of 8 patients, the average education size was more than 5 *cm*, and in two patients a malignant chemodectoma was revealed according to the results of histology.

Currently, the Shamblin classification system, which is based on the degree of involvement of the adjacent carotid arteries, is used to assess the risk of intraoperative bleeding and damage to cranial nerves and can indicate the potential need for resection of the carotid artery with revascularization [*Kim G et al., 2017*].

Also in the literature is the work of Kim G et al, who offer new predictors of complications of CBT resection. The study is based on measuring tumor volume and distance from the base of the skull (DTBOS) in conjunction with the Shamblin score [*Kim G et al., 2017*]. According to the results of the study, each 1 *cm* decrease in distance to base of skull is associated with a 1.8-fold increase in the risk of blood loss> 250 *ml* by a factor of 1.8 (CI 95%, 1.25-2.55) and a 1.5-fold increase in the risk of cranial nerve damage (CI 95%, 1.19-1.92). In our study, the assessment was carried out only according to the Shamblin classification.

The prolonged existence of even a benign tumor poses a threat of stenosis of the larynx, trachea, esophagus, involvement of the cranial nerves and great vessels of the neck with impaired cerebral circulation, and its spread into the cranial cavity. Surgical resection is recommended for all CBT due to the risk of local complications associated with mass size and malignancy.

Conclusion

Surgical resection is the treatment of choice for carotid body tumors. The large size of the masses and involvement of the carotid arteries and cranial nerves in the process directly affect the surgical approach and increase the risk of complications. Thorough preparing for surgery, mainly in patients with type III by Shamblin grade can significantly reduce major complications. In case of detection of malignant cells in a tumor or neighboring lymph nodes, it is recommended that an oncologist is monitored immediately after surgery for the possibility of radiation or chemotherapy. For patients who underwent excision of the tumor with replacement of the carotid artery it is recommended to perform ultrasound for the first 48 hours, as well as monitoring the patient's neurological status after waking up.

The rare occurrence, slow growing, asymptomatic course and a limited amount of information on the detection and management of the disease lead to an underestimation of the condition [*Makeieff M et al.,2008*].

REFERENCES

1. *Bemard RP* (1992). Carotid body tumors. Am J Surg 163(5):494-6. doi: 10.1016/0002-9610(92)90395-8
2. *Bougrine F, Maamouri F, Doghri R, Msakni I, Sabbegh Znaidi N, Chouchane O, et al* (2008). Une tumeur rare du glomus carotidien. J Mal Vasc 33(4-5):214-7. doi: 10.1016/j.jmv.2008.06.003
3. *Byrne JJ* (1958). Carotide body and allied tumor. Am J Surg 95(3):371-84. doi: 10.1016/0002-9610(58)90537-3.
4. *Céruse P, Cosmidis A, Belot A, Rabilloud M, Fuchsmann C, Poupart M, et al., (2014*). A pyriform sinus cancer organ preservation strategy comprising induction chemotherapy with docetaxel, cisplatin, and 5-fluorouracil, followed by potentiated radiotherapy. Anti-Cancer Drugs, 25(8), 970–975. doi:10.1097/CAD.0000000000000126 Anti-Cancer Drugs 25: 970-975, No. 8, Sep 2014 - France 803112869
5. *Daudi FA, Thurston OG* (1989). Carotid body tumours: the University of Alberta Hospital experience. Can J Surg;32(3):159-61
6. *Green JD, Olsen KD, DeSanto LW, Scheithauer BW* (1998). Neoplasms of the vagus nerve. Laryngoscope. 98(6 Pt 1):648-54. doi: 10.1288/00005537-198806000-00014.
7. *Kim GY, Lawrence PF, Moridzadeh RS, Zimmerman K, Munoz A, Luna-Ortiz K. et al* (2017*).* New predictors of complications in carotid body tumor resection. Journal of Vascular Surgery 65(6):1673-1679. doi: 10.1016/j.jvs.2016.12.124.

8. *Lack EE, Cubilla AL, Woodruff JM, Farr HW* (1977). Paragangliomas of the head and neck region: a clinical study of 69 patients Cancer 39(2):397-409. doi: 10.1002/1097-0142(197702)39:2<397::aid-cncr2820390205>3.0.co;2-c
9. *Lahey FM, Warren KW* (1947). Tumor of carotid body. Surg Gynecol Obstet.85(3):281–288
10. *Li J, Wang S, Zee C, Yang J, Chen W, Zhuang W, et al* (2010). Preoperative angiography and transarterial embolization in the management of carotid body tumor: a single-center, 10-year experience. Neurosurgery 67(4):941-948; discussion 948. doi: 10.1227/NEU.0b013e3181eda61d.
11. *Lim JY, Kim J, Kim SH, Lee S, Lim YC, Kim JW, et al* (2010). Surgical treatment of carotid body paragangliomas: outcomes and complications according to the Shamblin classification. Clin Exp Otorhinolaryngol 3(2):91-5. doi: 10.3342/ceo.2010.3.2.91.
12. *Makeieff M, Raingeard I, Alric P, Bonafe A, Guerrier B, Marty-Ane C* (2008). Surgical management of carotid body tumors. Ann Surg Oncol 15(8):2180-6. doi: 10.1245/s10434-008-9977-z.
13. *McPhersen GA, Hallyday AW, Mansfield AO* (1989). Carotid body tumor and other cervical paragangliomas: diagnosis and management in 25 pations. Br J Surg 76(1):33-36. doi: 10.1002/bjs.1800760111.
14. *Mitchell RO, Richardson JD, Lamberth GE* (1996). Charecteristics, surgical management and outcome in 17 carotid body tumor. Am Surg.62(12):1034–1037
15. *Muhm M, Polterauer P, Gstottner W, Temmel A, Richling B, Undt G, et al* (1997). Diagnostic and therapeutic approaches to carotid body tumors. Review of 24 patients. Arch Surg 132(3):279-284. doi: 10.1001/archsurg.1997.01430270065013.
16. *Gerecer-Gil NY, Figuera LE, Llamas FJ, Lara M, Escamilla JG, Ramos R et al* (2010). Mutation of SDHB is a Cause of Hypoxia-Related High-Altitude Paraganglioma. Clin. Cancer. Res. 15;16(16):4148-4154. doi: 10.1158/1078-0432.CCR-10-0637.
17. *Nora JD, Hallett JW, O'Brien PC, Naessens JM, Cherry KJ, Pairolero PC* (1988). Surgical resection of carotid body tumors: long-term survival, recurrence, and metastasis. Mayo Clin Proc 63(4):348-352. doi: 10.1016/s0025-6196(12)64856-3.
18. *Parry DM, Li FP, Strong LC, Carney JA, Schottenfield D, Reimer RR, et al* (1982). Carotid body tumors in humans: genetics and epidemiology. J Natl Cancer Inst. 68(4):573–578
19. *Power AH, Bower TC, Kasperbauer J, Link MJ, Oderich G, Cloft H, et al* (2012). Impact of Preoperative Embolization on Outcomes of Carotid Body Tumor resections. J. Vasc. Surg. 56(4):979-989. doi: 10.1016/j.jvs.2012.03.037.
20. *Pryse-Davies J, Dawson IM, Westbury G* (1964). Some morphologic, histochemical and chemical observations on chemodectomas and the normal carotid body, including a study of the chromaffin reaction and possible ganglion cell elements. Cancer 17:185-201. doi: 10.1002/1097-0142(196402)17:2<185::aid-cncr2820170208>3.0.co;2-1.
21. *Robison JG, Shagets FW, Becket WC, Spies JB* (1986). A multidisciplinary approach to reducing morbidity and operative blood loss during resection carotid body tumor. Surg Gynicol Obstet 168(2):166–170
22. *Scudder CL* (1903). Tumor of the intercarotid body. A report of one case, together with all the cases in the literature. Am J Med Sci.126(3)384–389
23. *Shamblin WR, ReMine WH, Sheps SG, Harrison EG Jr* (1971). Carotid body tumor (chemodectoma): clinicopathologic analysis of ninety cases. Am J Surg 122(6):732-739. doi: 10.1016/0002-9610(71)90436-3.
24. *Wang SJ, Wang MB, Barauskas TM, Calcaterra TC* (2000). Surgical management of carotid body tumors. Otolaryngol Head Neck Surg 123(3):202-206. doi: 10.1067/mhn.2000.106709.
25. *Ward PH, Jenkins HA, Hanafee WN* (1978). Diagnosis and treatment of carotid body tumors. Ann Otol Rhinol Laryngol 87(5 Pt 1):614-621. doi: 10.1177/000348947808700503.
26. *Werter IM, Rustemeijer C* (2013). Head and neck paragangliomas. Neth J Med.71(10):508-511.
27. *Williams MD, Phillips MJ, Nelson WR, Rainer WG* (1992). Carotid body tumor. Arch Surg 127(8):963-7; discussion 967-968. doi: 10.1001/archsurg.1992.01420080097016.
28. *Woolen S, Gemmete JJ* (2016). Paragangliomas of the head and neck. Neuroimaging Clin N Am. 26(2):259-278. doi: 10.1016/j.nic.2015.12.005.

THE NEW ARMENIAN MEDICAL JOURNAL
Vol.16 (2022). No 1

CONTENTS

THE NEW ARMENIAN MEDICAL JOURNAL

Vol.16 (2022). No 1

The Journal is founded by
Yerevan State Medical
University after M. Heratsi.

Rector of YSMU

Armen A. Muradyan

Address for correspondence:

Yerevan State Medical University
2 Koryun Street, Yerevan 0025,
Republic of Armenia

Phones:

(+37410) 582532 YSMU
(+37410) 580840 Editor-in-Chief

Fax: (+37410) 582532

E-mail: namj.ysmu@gmail.com, ysmiu@mail.ru

URL: http//www.ysmu.am

Our journal is registered in the databases of Scopus, EBSCO and Thomson Reuters (in the registration process)

SCOPUS

EBSCO

THOMSON REUTERS

Copy editor: Tatevik R. Movsisyan

Printed in "VARM" LLC
Director: Ruzanna Arakelyan
Armenia, 0018, Yerevan,
Tigran Mec 48, 43
Phone: (+374 91) 19 29 00,
E-mail: armana6@mail.ru